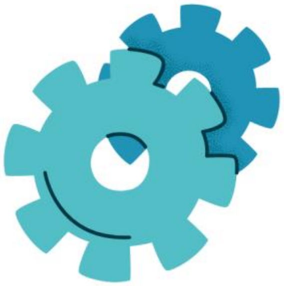
hdreach
Helping families with Huntington's disease

In-Home Care Training Guide for Huntington's Disease

A workbook for care partners caring for someone with Huntington's Disease.

This workbook has been created as a guide to help new-to-HD care partners understand the unique challenges caused by the disease. This workbook provides an overview of the disease, its stages, most common symptoms, and ways to navigate those symptoms.



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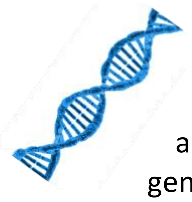
What is Huntington's Disease?



Huntington's Disease (HD) is a rare genetic **neurodegenerative** disorder that is passed down from one generation to the next. Each child of a parent with HD has a **50/50** chance of inheriting the disease. A genetic test administered after a person turns 18 can determine if the person has the genetic defect. HD symptoms typically start between the ages of **35-50** and gradually get worse over time. HD only affects the **brain**, so other organs such as the heart, kidneys, lungs, and liver are not directly impacted. In comparison with Alzheimer's disease or Parkinson's disease, persons with HD are usually younger and have fewer medical problems besides the neurological disease. There are **five defined stages** of HD. Some people transition quickly between stages and others progress slowly.

What are the Stages of HD?

Prodromal Phase (7-10 years before motor symptoms begin)

Subtle and gradual changes in thinking and behavior may become noticeable during this stage, but motor symptoms do not. Behavioral and medical treatments for these changes are available.

Stages 1 and 2 (Early HD)

During this stage, the person with HD has normal or mildly reduced work ability and might need some help with finances. A person with HD can still live independently and typically experiences minimal impairment in activities and chores. Psychiatric symptoms are intermittent and manageable and emerging motor symptoms might become more identifiable.

Stage 3 (Mid-Stage HD)

During this stage, walking, eating, and communicating become affected. There may be some noticeable impairment as it relates to money management and cognitive symptoms become less treatable than they were in the earlier stages. Frustrations easily mount and a person with HD is typically at a high risk of suicide during this time.

Stages 4 and 5 (Late-Stage HD)

During the final stages of Huntington's disease, all areas of functioning become severely impaired. At this stage, total care is often needed, and many enter long-term care facilities. Chorea can be severe, or movements can become rigid or stiff.

What are the Symptoms of HD?

Some symptoms you can see or observe easily, like the involuntary movements called chorea. Or you might hear slurred speech. Other symptoms are a little harder to identify, such as depression or difficulty making decisions. Symptoms of HD are typically divided into three categories: **Physical, Cognitive, and Psychiatric.** In the following pages, we'll look at these three categories and ways to address these symptoms from a caregiver's perspective.

Physical Symptoms

CHOREA

Chorea is dance-like uncontrollable movements and is the most well-known symptom. These movements often cause clumsiness, balance issues, and poor coordination.

Caregiver tips:

- Increased stress or anxiety often make movements worse.
- People with HD are not usually trying to kick or strike a caregiver during personal care, but the chorea can make it appear that way.
- Padding sharp corners or furniture can help prevent bruising.
- People in middle and late stages of the disease are at increased risk for falling. Many use durable medical equipment such as walkers, wheelchairs, bedside commodes, shower chairs, or grab bars to reduce this risk.
- Medications help reduce these movements and it is usually taken between 2 and 3 times a day. The medications are called Xenazine (Tetrabenezine) or Austedo (Detretrabenezine).
- PT/OT can help with balance and strength during the early and middle stages of the disease. During late-stage HD the person is often unable to walk and needs total assistance with transfers.

SWALLOWING/SPEECH ISSUES

Swallowing becomes an issue during mid- and late-stage HD. People with HD often cough and choke because the mechanisms involved in this process become weakened. Speech can be slurred and difficult to understand.

Caregiver tips:

- Soft foods, thickened liquids, and increased calories are often suggested by speech therapists.
- Create a safe and distraction free environment for eating. Have the person with HD sit upright at the table rather than in a recliner in front of the TV.
- Encourage small bites and second swallows to clear food from mouth.
- Give plenty of time to finish a meal.

Cognitive Symptoms

SLOWER THINKING AND DIFFICULTY LEARNING NEW THINGS

With fewer brain cells working at their best, it takes longer to process and retrieve information. As a result, the person with HD may be slower to respond, slower to remember things, have trouble switching to a new topic, and be unable to wait- for anything.

Caregiver tips:

- ✓ Ask ONE question at a time and WAIT for a response. It can take more than 10 seconds to process your question, formulate an answer, and coordinate a verbal response. Be okay with silence after you ask a question.
- ✓ Using multiple choice or yes/no questions are generally easier than open ended ones.
- ✓ People with HD hate surprises so keeping to a predictable routine is easiest.
- ✓ People with HD can learn new things, but it will take much longer to retain the information. Routine is important for managing day-to-day events.
- ✓ Slower thinking is exhausting. The person with HD may stop mid-task and give up because of the effort it takes to remember the steps. For example, when dressing they may get as far as undergarments and then stop. They will need cueing to continue.

EXTREME IMPATIENCE

Caregiver tip:

- ✓ If the person with HD wants something, do not make them wait if at all possible- this is part of the disease and not their personality.

GETTING STUCK OR PERSEVERATION

Getting stuck on a thought, subject, or idea is very common in HD. The person with HD may ask you the same question over and over, even if you have already answered multiple times.

Caregiver tips:

- ✓ Distract the person or redirect them by changing the subject, moving to a different room, offering a favorite snack.
- ✓ Use the same phrase to remind them to stop doing something. Some suggestions are:
  - ⇒ *We are done with that*
  - ⇒ *We aren't going to talk about that*
  - ⇒ *Use the time-out hand signal and stop talking*
- ✓ Remember that this behavior is a symptom and not their personality

UNAWARENESS (ALSO CALLED ANOSOGNOSIA)

A common symptom of HD is lack of awareness of symptoms or problem behaviors. Often people with HD do not notice their movements, balance issues, choking, or mood issues.

Caregiver tips:

- There is no treatment for anosognosia, so the best approach is to be aware of the person's unawareness.
- Trying to prove to a person with HD they have symptoms is proven not to work and will often make the situation worse.

TROUBLE WITH FACIAL EXPRESSIONS

This symptom is twofold, people with HD have trouble reading someone's facial expression. They will have a tough time interpreting if someone is happy, scared, angry, or bored. At the same time, the muscles in the face of the person with HD weaken over time, and they have trouble showing emotion. They could be smiling on the inside but appear bored or disinterested.

Caregiver tip:

- Remembering that a person with HD might appear disinterested or grimace, but this is a symptom. You might have to ask how they are feeling and then wait for an answer.

IMPULSIVITY/DISINHIBITION

Impulsivity is acting without thinking. Disinhibition is not feeling embarrassed or ashamed. When these symptoms appear, some people with HD lose their temper, drink too much, steal, or have inappropriate sexual relations. This symptom can have extremely negative results for the person with HD and everyone around them.

Caregiver tip:

- These symptoms can be helped with medication from a psychiatrist.
- Try to stay calm and attend to the emotion being expressed by the person with HD. Their hurtful words and actions were likely because of the disease and not their personality.

Mood Symptoms

DEPRESSION

Depression is quite common in HD and largely due to the HD brain not working properly.

Caregiver tips:

- ✓ Medications from a psychiatrist are typically highly effective in reducing depression.
- ✓ Keep a consistent schedule and routine. Having structure can help.
- ✓ Exercise and time spent outdoors/in the sun can have a positive impact on depression.

IRRITABILITY

Irritability in HD is common. The part of the brain that helps regulate emotions gets damaged and the result can lead to increased irritability. Sometimes it is not obvious what the goal or desired outcome of the outburst is.

Caregiver tips:

- ✓ Look for common triggers such as hunger, thirst, pain, exhaustion, change in routine, grief, unplanned events or surprises, or medication changes. Often when these triggers are addressed the irritability is greatly reduced.
- ✓ We can't always control the situation, but we can control how we react. If we react calmly, use a soft voice, and let the person with HD do most of the talking, they will have an easier time calming down themselves.
- ✓ The good news is irritability is highly treatable by medications from a psychiatrist.
- ✓ Remembering that irritability is a symptom and not part of their personality.

Helpful Links and Further Reading

- HD Reach website:
  https://www.hdreach.org/signs-symptoms/

- CME4HD- FREE online accredited learning:
  https://huntingtonstudygroup.org/cme4hd-online/

- HDSA's Educational Guides:
  https://hdsa.org/find-help/community-social-support/hdsa-resource-library/

- Further Reading: Hurry Up and Wait! A Cognitive Care Companion Huntington's Disease in the Middle and More Advanced Years by Jimmy Pollard
  https://smile.amazon.com/Hurry-Up-Wait-James-Pollard/dp/B06XYTYKZ3